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Dilated right heart in a young lady with syncope: atrial septal defect masquerading as arrhythmogenic right ventricular cardiomyopathy

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A 30-year-old female presented with palpitations and syncope. Electrocardiogram (ECG) showed sinus rhythm with abnormal T wave inversions in the precordial leads (Figure 1*A*). A 2D echo revealed a dilated right ventricle (RV), mid-RV dimension of 4.0 cm (normal 1.9–3.5 cm). Cardiac magnetic resonance imaging (CMR) was suspicious for arrhythmogenic right ventricular cardiomyopathy

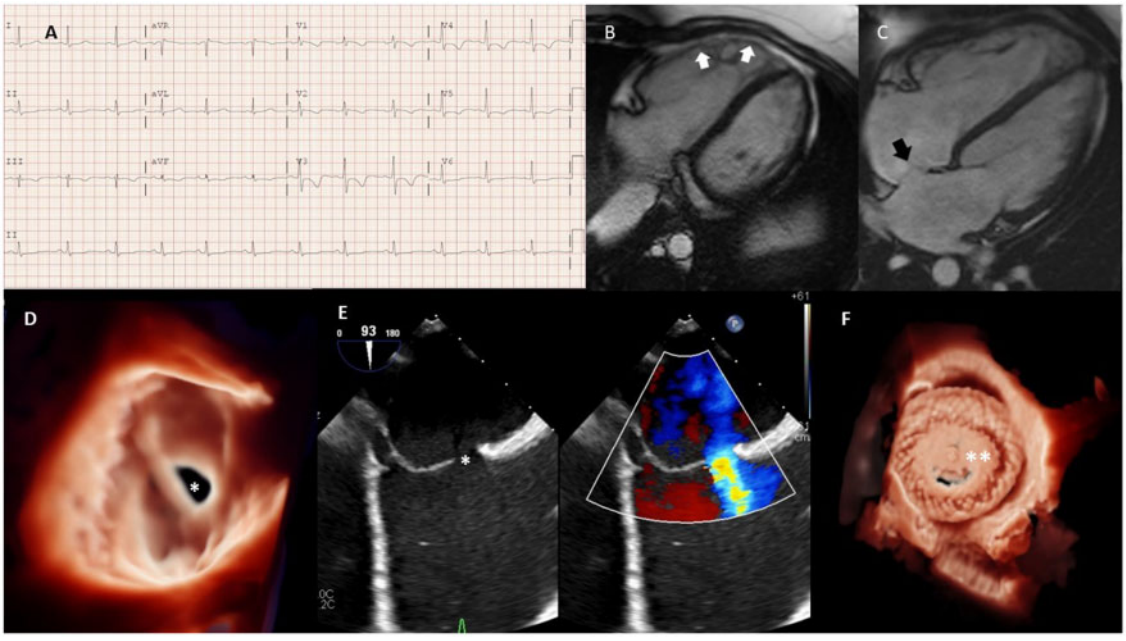

Figure 1 (A) Sinus rhythm with T wave inversions in V1–V6. (*B* and *C*) Cardiac magnetic resonance imaging four-chamber view: white arrows—dilated right ventricle with suggestion of regional wall motion abnormalities; black arrow—hypermobile interatrial septum. (*D*) 3D rendered transillumination (Trueview) of the septum secundum atrial septal defect on transoesophageal echocardiogram. (*E*) Bi-caval view on transoesophageal echocardiogram demonstrates septum secundum atrial septal defect. (*F*) 3D rendered transillumination (Trueview) of the atrial septal defect closure device on transoesophageal echocardiogram.

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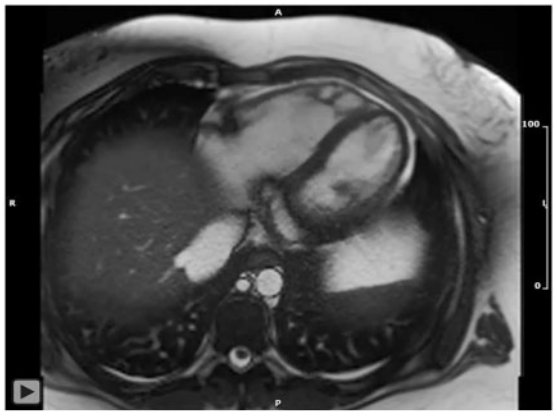
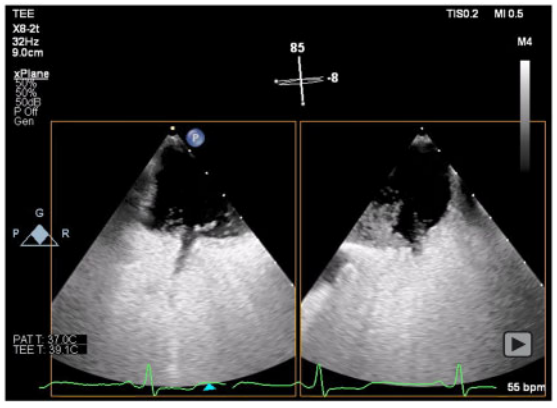
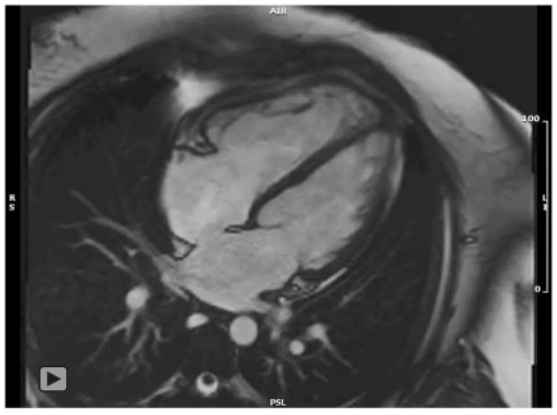


Video 1 Cardiac magnetic resonance imaging cine images; right ventricular dilation with suggestion of regional dyskinesia of the right ventricle free wall.



Video 3 Bi-caval view on transoesophageal echocardiogram that shows the septum secundum atrial septal defect. Injection of agitated saline during the transoesophageal echocardiogram reveals the bidirectional nature of the shunt with predominant left to right shunt.



Video 2 Cardiac magnetic resonance imaging cine images; four-chamber view that reveals the hypermobile interatrial septum without an obvious atrial septal defect.

(ARVC) with RV dilation and a suggestion for regional wall motion abnormalities (RWMA) (*Figure 1B* and *Video 1*). No late gadolinium enhancement or fibro fatty infiltration of the RV was noted; the right ventricular ejection fraction (RVEF) was calculated at 55%. The right atrium (RA) was abnormally enlarged with hypermobile interatrial septum (IAS) (*Figure 1C* and *Video 2*). The history of syncope, abnormal T waves on ECG, and RV dilation with dyskinesia on CMR were concerning for ARVC. However, the abnormally dilated RA and RV with a hypermobile IAS prompted us to perform a 3D transoesophageal echocardiogram. This revealed an ostium secundum atrial septal defect (ASD) with significant left to right shunt (* in *Figure 1D* and *E* and *Video 3*). On re-review, no RWMA were observed. In fact, the abnormalities were considered to be caused by volume overload due to ASD. The RV findings were thus likely an 'ARVC mimic' secondary to the ASD. Her ASD was closed with a percutaneous closure device (** in *Figure 1F*). Her initial symptoms of palpitations and syncope were further evaluated with a 30-day event monitor that revealed occasional premature ventricular contractions that could explain her palpitations. No sustained arrhythmias were noted. She did not have recurrent syncopal events.

Her baseline 2D transthoracic echo did not have a good subcostal echocardiographic window; which would have been ideal to capture the ASD (Supplementary material online, *Video S1*). CMR is an ideal test to evaluate for structural cardiac anomalies. However, the ASD was not obvious on CMR imaging (Supplementary material online, *Video S2*). This goes on to highlight the importance of multimodality imaging in this particular case.

The diagnosis of ARVC is complicated and patients referred for CMR for suspected ARVC can have an alternate diagnosis. A detailed history, clinical assessment with multimodality imaging approach is critical. A deliberate effort has to be made to rule out other conditions that mimic ARVC phenotypes. This has a major impact in the clinical management.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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